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Case Report

Fetal intra-abdominal umbilical vein varix: a rare perinatal diagnosis

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ABSTRACT

Fetal intra-abdominal umbilical vein varix (FIUVV) is a rare perinatal diagnosis. It can be missed easily on routine 2D ultrasound. It is focal dilatation of umbilical veins diagnosed prenatally on ultrasound. we are reporting here the case of FIUVV this case to raise awareness about the FIUVV as it can be missed easily on routine ultrasound scan. A 31-year-old lady presented in her third pregnancy. She has had two previous uncomplicated normal vaginal births at term. Her scan at 31 weeks of gestation demonstrated estimated fetal weight at 64th centile, normal liquor volume and normal Doppler studies. A three-vessel umbilical cord and a small intra-abdominal umbilical vein varix of 12×15 mm. Further scan that was performed at 37 weeks of gestation showed increased in size of FIUVV of about 18×14 mm, however, no turbulence flow noted at the site of varix. Estimated fetal weight calculated as 2876 grams with normal Doppler studies and liquor volume. She has been advised delivery around 37-38 weeks. She is induced at 38 weeks of gestation followed by normal vaginal birth of an alive baby with APGAR scores of 9 at 1 minute, 10 at 5 minutes and 10 at 10 minutes of life. Baby initial checks didn't show any congenital abnormalities.

Keywords: Fetal, Umbilical, Foetuses, Pregnancy, Intra-abdominal

INTRODUCTION

Fetal intra-abdominal umbilical vein varix (FIUVV) is a focal dilatation of umbilical veins diagnosed perinatally on ultrasound.[1] It is also called intra-amniotic portion of the umbilical vein.

The incidence of FIUVV is 0.4-1.1/1000.[1-3] It is associated with congenital anomalies in about 43% of the foetuses resulting in intrauterine foetal death.[2] It is also associated with chromosomal anomalies is 2.8% of cases.[3,4,6]

CASE REPORT

A 31-year-old lady presented in her third pregnancy. She has had two previous uncomplicated normal vaginal births at term. Her course of index pregnancy was uneventful till 31 weeks of gestation when she was offered to have a growth scan due to suspected increased fundal height measurements during routine antenatal check-up. Her scan demonstrated single alive intrauterine pregnancy with marginal cord insertion and an anterior low placenta. Her booking blood results were unremarkable. vital signs were normal, blood pressure (BP) 108/73 mmHg, P: 81b/min, T 36.8). Her anomaly scan performed at 20 weeks of gestation was also unremarkable. However, this scan has ruled out low lying placenta. Her scan at 31 weeks of gestation demonstrated estimated fetal weight at 64th centile, normal liquor volume and normal Doppler studies. A three-vessel umbilical cord and a small intra-abdominal umbilical vein varix of 12×15 mm. Further scan that was performed at 37 weeks of gestation showed increased in size of FIUVV of about 18×14 mm, however, no turbulence flow noted at the site of varix. Estimated fetal weight calculated as 2876 grams with normal Doppler studies and liquor volume. She has been advised delivery around 37-38 weeks. She is induced at 38 weeks of gestation followed by normal vaginal birth of an alive baby

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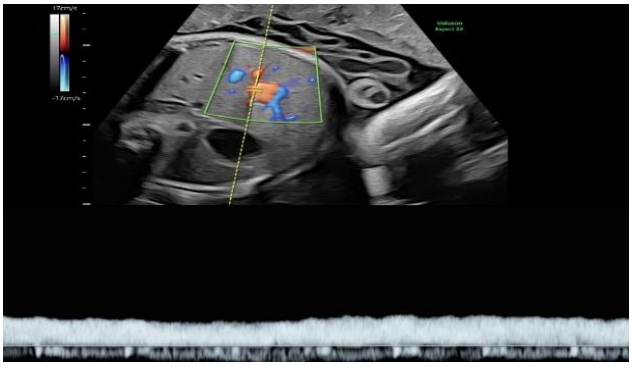

Figure 1: Doppler flow within the varix.

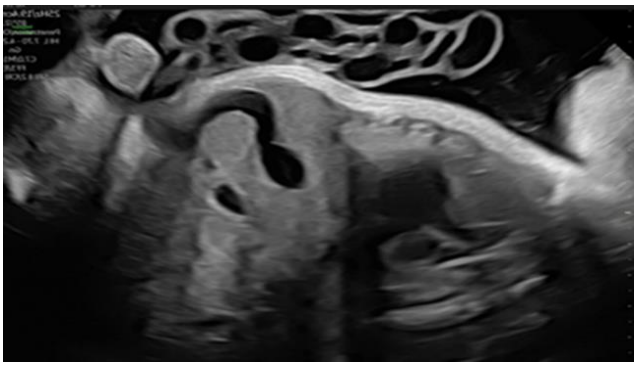

Figure 2: 2D image showing fetal intra-umbilical vein varix.

DISCUSSION

FIUVV is considered to be a developmental lesion rather than a congenital anomaly because more than two-thirds of the cases are diagnosed after 30 weeks of gestation. On ultrasound it can be seen as a round cystic structure between the inferior part of liver and anterior abdominal wall with venous flow within on color Doppler interrogation.[1,4] The diameter of intra-abdominal part of umbilical vein varies throughout the pregnancy ranging from 2-8 mm from early gestation till term.[3] The left umbilical vein enters the fetus at an umbilical ring and runs along the inferior border of the liver. It joins the portal system making the umbilical–portal system and enters the inferior vena cava through ducts venous. Inherent weakness or an unsupported intraabdominal portion of an umbilical vein makes it vulnerable to pressure changes, resulting in segmental dilatation of the vein. The diameter of umbilical vein increases linearly with gestational age, and the diagnosis is made when the diameter of umbilical vein exceeds two standard deviations for the gestation age.[2]

Various definitions of FIUVV are used in literature, most authors agreed on linear diameter of umbilical vein greater than 9mm or more than two standard deviations above the mean for gestational age or diameter of dilated part at least 1.5 times bigger than non-dilated part.[2] It is a diagnosis of exclusion. Other differential diagnosis includes intra-abdominal cysts including gall bladder cyst, choledochal or urachal cyst, cystic lymphangioma, enteric duplication cyst, rare persistent vitelline vein (VV) aneurysm resulting in postnatal thrombosis of the portal and systemic venous systems.[1,2] Literature shows more favorable outcomes if FIUVV is an isolated finding with significantly lower association with aneuploidy and mortality.[4,5,9]

The complications of FIUVV includes rupture, thrombosis, compression of the umbilical artery and other veins, and cardiac failure due to vascular sneaking by the varix causing rise in cardiac preload.[4]

Guidance on management is still not limited.[1,8] Regular fetal surveillances by fetal medicine unit every 2-3 weeks with umbilical artery dopplers should be offered to every woman diagnosed with FIUVV without the adjacent chromosomal/anatomical anomalies to prevent any complications specially Foetal anemia and thrombosis of umbilical vein resulting in superior mesenteric vein obstruction or portal hypertension as, it can lead to intrauterine fetal death.[5,7] Delivery should be recommended by 37-38 weeks either by labor induction or caesarean section depending upon obstetric indication.

CONCLUSION

FIUVV is rare ultrasound finding that can be missed easily and can be diagnosed by careful evaluation of the fetal umbilical vein after excluding other differential diagnosis. It needs careful fetal monitoring failing to do so can lead to the adverse outcome including intrauterine fetal death due to thrombosis of umbilical vein.

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